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Management of a Rare Case of Moyamoya Disease Associated With Left Posterior Cerebral Artery Aneurysm

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ABSTRACT

Background: *The association of moyamoya disease and posterior cerebral artery aneurysms is relatively rare and the prognosis is poor. The management is extremely challenging for neurosurgeons.*

Case Report: *We report a case of a 58-year-old man with a history of moyamoya disease who admitted complaining of sudden-onset headaches, vomiting, and seizure and the investigations revealed a subarachnoid hemorrhage, intracerebral hematoma, posterior cerebral artery aneurysms on a moyamoya disease field. The patient was treated surgically by the clipping of the aneurysm, but developed three days later a complication of the MMD and died.*

Conclusion: *The prognosis of this association is very poor, explain the fact that a therapeutic strategy should be done to reduce the mortality and to determine the best management which is surgery versus embolization and revascularization.*

Keywords

Moyamoya Disease, Aneurysm, PCA, DSA Angiography, Subarachnoid hemorrhage, Hematoma, Ventricles, Surgery management.

Abbreviations

CT: Computed tomography; DSA: Digital subtraction angiography; EDMS: encephaloduromyosynangiosis; EGS: encephalogaleosynangiosis; EMS: encephalomyosynangiosis; EVD: Extraventricular drainage; GCS: Glasgow Coma Scale; ICA: Internal carotid artery; ICU: Intensive care unit; MCA: Middle cerebral artery; MMD: Moyamoya disease; PCA: Posterior cerebral artery; Pcom: Posterior communicating artery; SAH: Subarachnoid hemorrhage; STA: Superficial temporal artery.

Introduction

Aneurysms of the posterior circulation are very challenging despite the development of microsurgical techniques in vascular surgery. The association of these aneurysms with a Moyamoya disease (MMD) is rare and the incidence is estimated to be 3-14% in adults [1].

MMD is a cerebrovascular disease uncommonly causing progressive stenosis od occlusion of the supraclinoid internal carotid artery (ICA) and its main branches within the Willis Circle [2,3]. MMD is extremely rare in Morocco and the prognosis is relatively poor.

The aneurysms could appear in the peripheral arteries or the major arteries [4]. The main trunk aneurysms, are formed due to hemodynamic disturbance, and have a higher risk of spontaneous progression or rupture, resulting in death. Here, we present a case of a patient who presented a posterior cerebral artery aneurysm associated with an MMD located in the main trunk. The aneurysm was well clipped but the patient presented others complications of the disease and died. The rare combination and the management of these cases are discussed in this paper.

Case Presentation

A 58-year-old man, with no medical history except chronic smoking.

In 2018, he presented headaches for one month followed by left eye ptosis. Physical examination showed no motor or sensitive deficit and III nerve complete palsy.

He performs a head Computed Tomography (CT) scan followed by a Digital Subtraction angiography (DSA) showing a bilateral Internal Carotid Artery stenosis an aspect of a Moyamoya disease (MMD). Stenosis of all ICA left and right and puff smoke peripherical arteries (Figure 1).

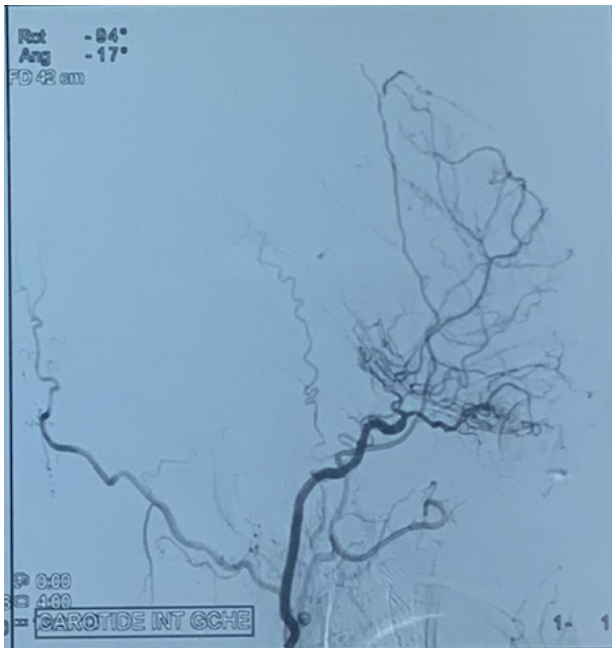

Figure 1: Digital subtraction angiography showing left stenosed ICA providing only peripherical arteries and aspect of Puff Smoke arteries.

The vertebral artery angiography revealed that all the vascularization was made by the left vertebral artery which is very massively developed and supplied the anterior circulation through the posterior communicating artery. Also, there is a left posterior cerebral artery thrombosed aneurysm (Figure 2). The patient was put on drugs and he has been released from the hospital and followed by a neurologist.

In July 2021, the patient comes back but this time in a neurosurgical emergency. He presents headaches followed by vomiting with grand mal seizures. On clinical evaluation, he was Glasgow Coma Scale (GCS) 14, with a stiff neck, meningeal syndrome, and right 4/5 hemiparesis. The III nerve palsy was partial, just small ptosis.

A head CT scan was performed showing a mesencephalic hematoma with subarachnoid hemorrhage (SAH) and intraventricular hemorrhage with hydrocephalus a Fisher IV SAH [Figure 3]. Extraventricular drainage (EVD) was performed in an emergency to reduce intracranial hypertension and treating the hydrocephalus and also ventricular hemorrhage.

Furthermore, he performed a DSA that showed a right posterior cerebral aneurysm, P1 segment, just before the origin of the right posterior communicating artery with dimensions of 3,03 x 2,35 x 5,65 mm and the neck was 4,81 mm (Figure 4). Also, the thrombosed left distal PCA.

The patient has cerebral vascularization only made by the vertebral circulation.

Our neuroradiologist cannot try the embolization because of the large size of the neck of the aneurysm. He waited two weeks, to reduce the risk of vasospasm, and also to discuss and planned the right management of this case. The EVD was removed after the cleaning of the ventricles and regression of the hydrocephalus.

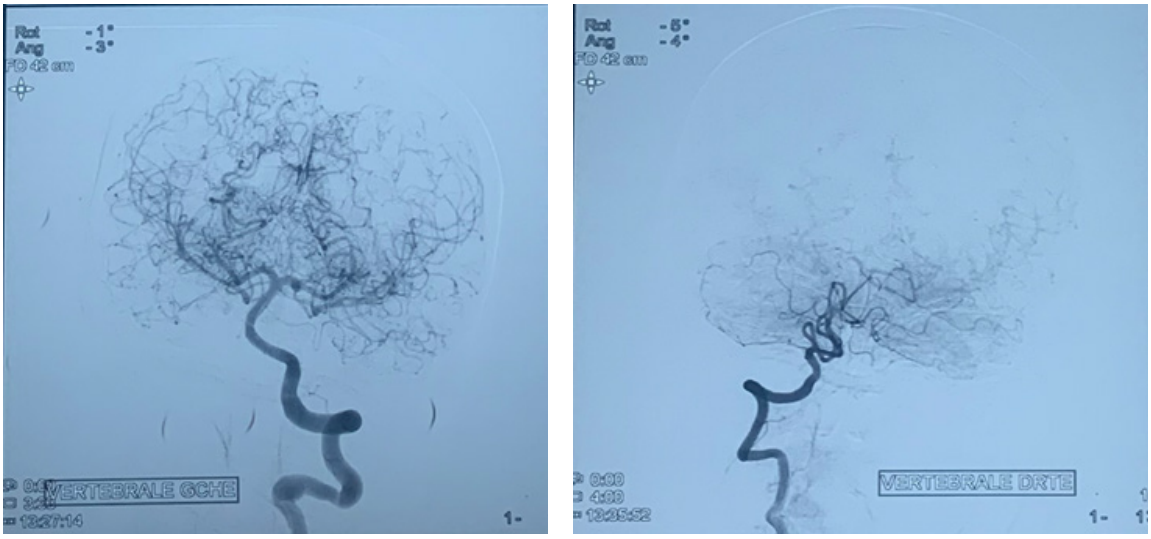

Figure 2: Digital subtraction angiography showing Vertebral arteries. (a): Left vertebral artery is very developed providing right and left PCA. (b): Right vertebral artery which is smaller but normal caliber.

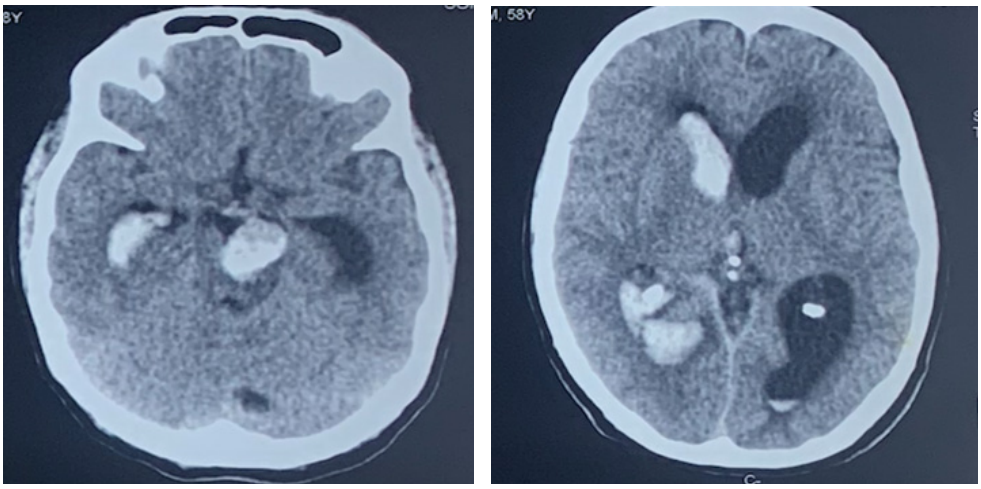

Figure 3: Head Computed Tomography, axial views. (**a**): Mesencephalic hematoma with dilatation of temporal horns with hematoma. (**b**): Hydrocephalus in lateral ventricles with hemorrhage.

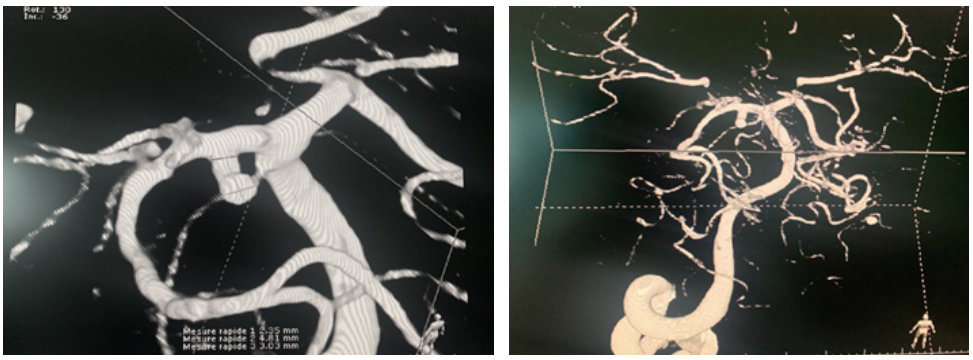

Figure 4: Second Digital subtraction angiography three-dimensional views (3D). (**a**): Profile view and a surgical view showing a left PCA aneurysm with measures. (**b**): Front view showing the left PCA aneurysm just before the origin of the Pcom artery.

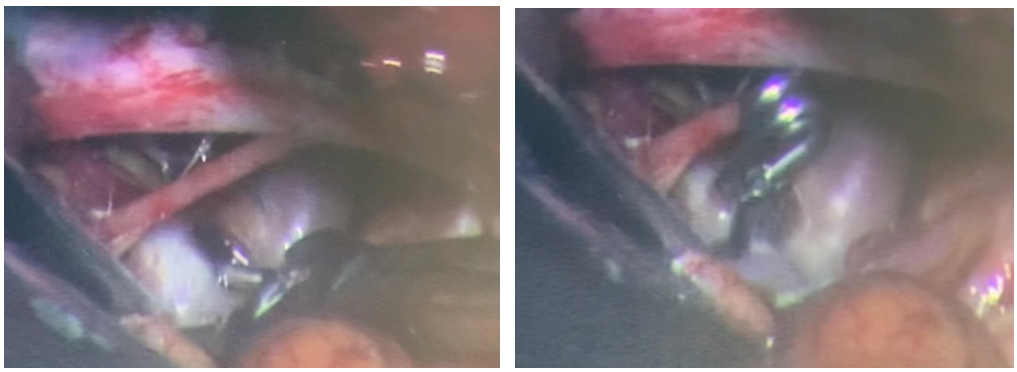

Figure 5: Operative view. (**a**) After sub-temporal approach, and temporal lobe spacing, visualization of the III nerve, then opening of Liliequist membrane, visualization of the basilar artery and the left PCA and the aneurysm which is dissected, and a right clip of 11mm was used. (**b**): After the aneurysm was clipped, verification of the permeability of the basilar artery and the PCA artery. The head of the clip was stuck on the third nerve, so we put a Surgicel between them to limit the III nerve palsy.

The surgical risk is higher than with common SAH because of the potential reduction in cerebral hemodynamic reserve, so our attention should be concentrated on the planning, the risk of pre- and postoperative vasospasm, and the timing of surgery.

After planning and discussion, we decided to approach firstly the aneurysm by clipping and secondary try the revascularization and that will be our first-time intracerebral bypass. Concerning the surgery, we chose the left sub-temporal approach.

During the surgery, the ICA was very thick and presented two thin arteries on his posterior. After seeing the III nerve, the basilar artery was found, and also the left PCA. The aneurysm was found dissected and clipped with a right clip of 11mm (Figure 5).

After surgery, the patient has stayed in intensive unit care (ICU) for 24 hours and return to the neurosurgery unit. He has remnant hemiparesis with a III nerve total palsy. 03 days after the surgery, he develops right hemiplegia. A CT scan was performed showing an insular hematoma and hydrocephalus (Figure 6). He started presenting trouble of consciousness with a Glasgow of 10. We went for a new EVD and he has been admitted to ICU. Unfortunately, he passed away only 2 days after his admission to ICU.

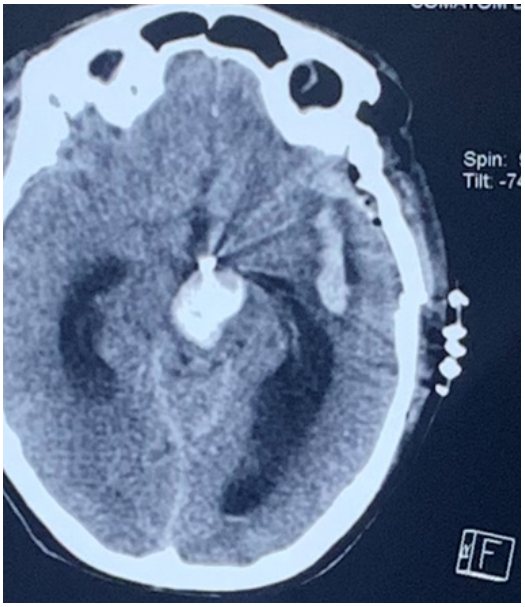

Figure 6: Postoperative head CT scan, axial view, showing left subtemporal craniotomy the mesencephalic hematoma and the clip, and also an intracerebral hematoma in the Sylvian fissure insula area with hydrocephalus.

Discussion

MMD is extremely rare in the African population and Morocco. A few case series have been reported in the literature, but the incidence and prevalence have not been done yet for this continent. Indeed, the epidemiology of moyamoya disease remains still unknown.

However, the diagnosis is based on angiographic characteristics such as stenosis affecting the distal internal carotid artery and/or proximal circle of Willis's vessels with the presence of prominent basal collateral vessels [5]. In this case, the DSA indicated that there was a bilateral occlusion of the ICA at the skull base with collaterals vessels abnormal in the related arteries that confirmed the diagnosis of MMD.

The incidence of aneurysms associated with MMD is 1.5 to 12.9% [6]. MMD is known to be associated with posterior cerebral aneurysms [3]. Physiopathology is that the remodeling is the increase of the blood flow through the vertebral artery to maintain the entire cerebral circulation. This leads to hemodynamic stress on the vertebral arteries, so in the basilar artery bifurcations causing aneurysms.

The medical literature on the surgical management of MMD-associated with aneurysms is limited to case reports and small series, the results of which suggest that direct surgical intervention generally results in good outcomes [7].

Therapeutic Strategy is not well established in the literature. There are no effective medical therapies for moyamoya disease. Through the provision of collateral pathways, surgical revascularization is the most successful therapy to improve cerebral hemodynamics and to reduce the risk of stroke and bleedings [8,9].

As far as aneurysms of posterior circulation associated with moyamoya disease are concerned, coiling combined with revascularization is the best option. In our case, the neck of the aneurysm was large and not a candidate for coiling embolization.

Surgical treatment of PCA aneurysms was described firstly by Hanafee and Janetta in 1966 [10]. And since then; many techniques and approaches have been used in the literature. For instance, in the P1 segment and P1-P2 segments, some authors preconized a pterional approach. And for the P2 and P3 segments of PCA, the subtemporal approach [11]. In this case, we choose the subtemporal approach because of the posterior orientation of the aneurysm and its relation with the posterior clinoid process. That was the best root to access the aneurysm even if located in the P1 segment of the PCA.

The revascularization procedures are well described in the literature [12,13]. Techniques described are the direct procedure which is bypass between superficial temporal artery to middle cerebral artery (STA-MCA) anastomosis, with indocyanine green fluorescence to access the bypass patency. And also the indirect procedure such as the encephalomyosynangiosis (EMS), encephaloduromyosynangiosis (EDMS), encephalogaleosynangiosis (EGS), and multiple burr hole surgery [14]. The STA, dura mater, temporal muscle, and galeal tissue are used as the pediculate donor tissues in these techniques. Indirect bypass surgery is technically simple to do and induces spontaneous angiogenesis between the brain surface and the vascularised donor tissues [15]. However, the beneficial effects are not immediate because surgical collaterals require 3-4 months to develop [16,17]. In our case, our department has not experimented with bypass techniques, so the Indirect Bypass would be easier and effective for tour case-patients. Our strategy was to firstly the treatment of the aneurysm by microsurgical clipping followed by the indirect bypass. Unfortunately, the patient developed another vascular complication of the MMD and died few days after surgery.

The prognosis of MMD associated with the aneurysm is relatively poor. The mortality rate reported in the Jian Yan series in 2019 about 104 cases of patients, was around 27,16%. For patients who had conservative treatment, death occurred in 47% of cases and for patients who had surgery, death occurred in 13% of cases [18]. That explains how poor is the prognosis and surgery must be considered for those patients to reduce mortality and to prevent patients from rebleeding.

Conclusion

The management of MMD associated with PCA aneurysms is very challenging. Our rare case represented the opportunity to discuss the surgical management of such aneurysms and the surgical treatment of MMD. The therapeutic that result of this case is the treatment of the aneurysm such as embolization, if possible of microsurgical clipping, and the revascularization (direct or indirect bypass) of MMD at the same time to prevent rebleeding, stroke and reduce the mortality rate.

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